Thirteen and Half Syndrome in Sero-negative Neuromyelitis Optica Spectrum disorder: A Case Report

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Abstract

Thirteen and a half syndrome is one and a half syndrome along with ipsilateral facial, trigeminal nerve lower motor neuron palsy . We describe a 33 year old man ,who developed acute onset right facial , trigeminal nerve palsy along with one and a half syndrome, thus constituting thirteen and a half syndrome , with sensory symptoms in left upper limb. MRI Brain showed T2 hyperintense and T1 hypointense lesion in right half of pons and cervicomedullary junction which was non enhancing after contrast administration. Serum antiaquaporin 4 antibodies were negative. He was diagnosed with sero-negative NMOSD .Patient improved significantly after 5 cycles of plasmapheresis Only 2 cases of thirteen and half syndrome have been reported in literature so far.

Keywords- One and half syndrome, Thirteen and half syndrome, Neuromyelitis optica spectrum disorder, Aquaporin-4 antibodies , Medial longitudinal fasciculus , pontine paramedian reticular formation

INTRODUCTION-

One-and-half syndrome is caused by a lesion in pontine tegmentum on one side , causing damage to Medial longitudinal fasciculus and pontine paramedian reticular formation.Thirteen and a half syndrome is characterised by ipsilateral facial ,trigeminal nerve lower motor neuron palsy and same sided one and a half syndrome

Case report :

We report a case of 33 year old male with no comorbidities ,presented with acute onset right facial weakness , numbness over right half of face and double vision since 10 days .Double vision was more on right lateral gaze .
Examination revealed inability to adduct right eye and nystagmoid movements in left eye on lateral gaze thus constituting right inter-nuclear ophthalmoplegia. Conjugate horizontal gaze palsy was present on right side . Sensation of light touch , pin prick in distribution of right trigeminal nerve(all three divisions) were impaired and corneal reflex absent on right side . He also had right facial nerve lower motor neuron palsy

He had a past history of similar such complaints (diplopia ,tingling numbness in left upper limb) 4 months back lasting for a week for which he was not evaluated and recovered without any treatment. Initial haematological studies were unremarkable. Cerebrospinal fluid study was normal. Erythrocyte sedimentation rate, C-reactive protein normal , Veneral disease research laboratory ,Serum myelin oligodendrocyte glycoprotein ,CSF and Serum oligoclonal bands , Anti-aquaporin-4 antibodies (IgG) ,Antinuclear antibodies (ANA) by Immunofixation , ANA blot . p-Antineutrophil cytoplasmic antibodies(p-ANCA) & c-Antineutrophil cytoplasmic antibody (c-ANCA)were all negative.

Magnetic resonance imaging study revealed T2 and FLAIR hyperintense lesion in right side pons extending into right cerebellar peduncles ;lesions appeared hypointense on T1 image and did not show any contrast enhancement . No diffusion restriction was noted. MRI spine study revealed T2 hyperintense signal in cervicomedullary junction .MR spectroscopy revealed no abnormal findings .Positron emission tomography (PET-CT) scan did not reveal any abnormal FDG uptake
He was treated as seronegative Neuromyelitis optica spectrum disorder with intravenous methylprednisolone 1 g daily for 5 days with no significant improvement .Following this 5 cycles of plasmapheresis were done .Patient showed good response with return of sensations on face, improvement in facial weakness and ocular movements. Patient was followed up on maintenance dose of steroids with no recurrence of symptoms over a period of six months .

DISCUSSION:

One-and-half syndrome is caused by a lesion of pontine tegmentum on one side causing damage to the pontine paramedian reticular formation and Medial longitudinal fasciculus. It was First reported and named by Fisher in 1967. It is characterised by ipsilateral internuclear ophthalmoplegia (INO) and ipsilateral conjugate horizontal gaze palsy[1]. A series of related rare syndromes called one-and-half syndrome spectrum disorders has been named after one and half syndrome [2] The diagnosis of one-and-half syndrome and its spectrum disorders relies on cranial nerves involved ,clinical symptoms and MRI findings[3]. The treatment varies according to etiology. Only 2 cases of thirteen and half syndrome have been reported [1,2] so far. Common causes for one-and –half syndrome were cerebrovascular disease , demyelination followed by infectious causes[4]. Less common causes were head injury, brain stem tumor [5]

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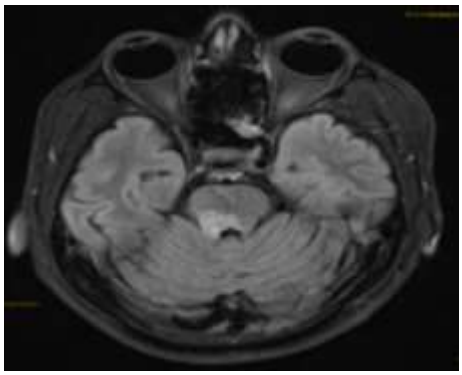
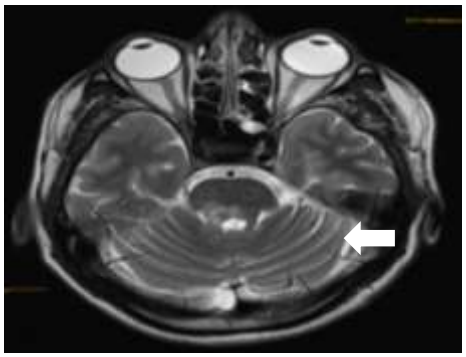
FIGURE-1:



(a)



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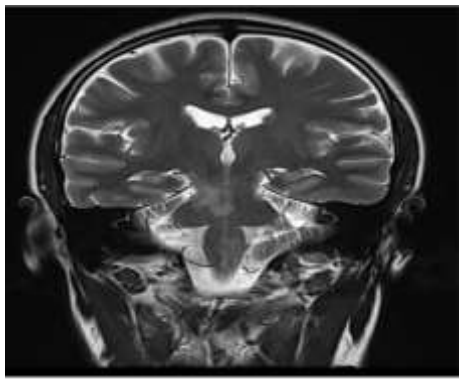
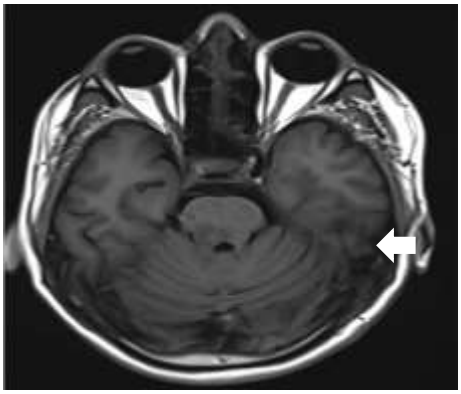
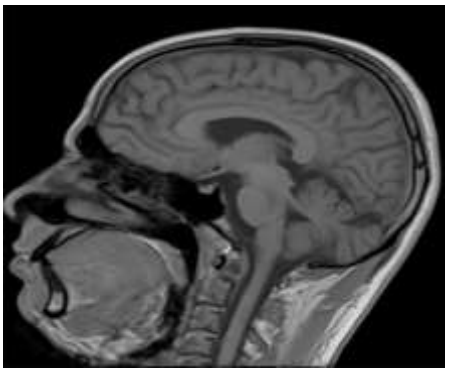


(c)



(d)



(e)

Figure -1 showing Axial flair (a) and axial T2 (b) and coronal T2 (c) showing hyperintensity within the antero lateral aspect of right side of pons ,figure (d) showing Axial T1 and sagittal T1W (e) showing hypointense areas in anterolateral pons(white arrows)

FIGURE -2 :

(a)

(b)

(c)

(d)

FIGURE 2 (a) and (b) showing LMN right 7 cranial nerve PALSY before plasmapheresis
Fig 2 (c) and (d) Showing improvement in LMN right 7 cranial nerve Palsy after plasmapheresis

FIGURE-3 :

(A)

(B)

(C)

(D)

Figure-3 (A) (B) showing eye position in neutral, left gaze respectively before plasmapheresis.
(C)& (D) showing eye position in neutral, left gaze respectively after plasmapheresis.